Fetal Ventricular Pacing for Hydrops Secondary to Complete Atrioventricular Block

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The advent of ultrasound recording has expanded the capabilities for treatment of the fetus in utero. The diagnosis of specific disease processes has allowed for prenatal intervention by new techniques designed to improve fetal survival. The application of ventricular pacing in a hydropic fetus with complete atrioventricular (AV) block is reported. Complete AV block resulted from maternal collagen vascular disease. The application of ventricular pacing was to allow for further in utero development and for reversal of hydrops fetalis after improvement in cardiac output. Despite fetal death 4 hours after placement of the ventricular pacing lead, this procedure when applied earlier in the development of hydrops may allow for fetal survival.

Ventricular pacing was accomplished without apparent trauma to mother or fetus and no evidence of fetal injury was seen at necropsy. Therefore, in the fetus who would otherwise die in utero before the point of viability ex utero, fetal ventricular pacing may be a rational alternative to current observation.

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Complete atrioventricular (AV) block is usually well tolerated in utero during the third trimester unless the ventricular rate is extremely slow or associated cardiac defects result in AV valve regurgitation. Nonimmunologic hydrops fetalis may result (1,2), which represents severe fetal cardiac decompensation due to congestive heart failure. The hydropic fetus with complete AV block will die in utero if left untreated in almost all cases. At present, a viable hydropic fetus with complete AV block is delivered by cesarean section, and temporary ventricular pacing is initiated during perinatal resuscitation (3). However, even after this course, perinatal mortality exceeds 80% (3,4). Because the prognosis for the hydropic fetus is poor, we have applied a new technique of maternal transuterine-fetal transthoracic ventricular pacing.

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Case History

Maternal history. A 24 year old white woman, gravida II para 1-0-0-1, with Sjögren's syndrome was referred for fetal cardiac evaluation at 27½ weeks' gestational age. Sjögren's syndrome was diagnosed 1 year previously when she developed xerophthalmia and had an antinuclear antibody titer greater than 1:1,000. She was asymptomatic and required no medications. Normal fetal heart sounds were noted by Doppler auscultation until 20 weeks' gestation when fetal bradycardia at 55 beats/min was noted.

Medical management. At 24 weeks' gestation, an obstetrical ultrasound examination in another hospital revealed moderate polyhydramnios and fetal ascites. Fetal cardiac anatomy appeared normal. Treatment with terbutaline, 5 mg orally every 6 hours, was initiated to increase the fetal ventricular rate. At 26 weeks' gestation, a follow-up maternal ultrasound examination suggested depressed right ventricular contractility and right ventricular dilation. Bilateral pleural effusions developed. Intravenous digoxin, 1 mg, was administered over 24 hours and maintenance treatment with oral digoxin, 0.25 mg daily, was begun. She received intramuscularly 24 mg of betamethasone acetate

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and betamethasone sodium phosphate. Three days later the fetal ventricular rate was 44 beats/min. Intravenous atropine, 0.25 mg, and an isoproterenol infusion, 2.75 μg/min, resulted in maternal tachycardia without change in the fetal ventricular rate. Spontaneous fetal movement disappeared and the patient was transferred to this institution.

Fetal echocardiography. Two-dimensional echocardiographic evaluation (Acuson-128) revealed complete AV block with an atrial rate of 128/min and a ventricular rate of 32/min. A 3 mm pericardial effusion had developed. The left ventricular shortening fraction was 47% with an internal diastolic diameter of 10 mm. The right atrial wall was hypertrophied and the right ventricle was markedly enlarged (16 mm end-diastolic dimension), using the criteria of DeVore et al. (5). No structural defect was present. Pulsed Doppler sampling (ATL 600) in the right and left atria at the level of the AV valves revealed mild tricuspid and mitral insufficiency.

Fetal pacing procedures. Informed consent for transuterine transthoracic fetal ventricular pacing was obtained after extensive discussions with the physicians involved and a presentation to the parents of all known risks and potential benefits as well as an explanation of the uncertainties.

The mother was not sedated for the procedure. Xylocaine, 1%, was infiltrated subcutaneously in the maternal abdomen. The fetus remained quiet in a left occiput posterior position. Under continuous sector scanning, a 17 gauge Tuohy needle was inserted into the uterine cavity and was advanced into the fetal thorax and into the right ventricle (Fig. 1). Removal of the obturator yielded pulsatile blood flow. Although considered, no attempt was made to obtain cardiac blood for arterial blood gases or digoxin level determinations.

An Electro-Catheter Corporation model 11-KPJ1 bipolar pigtail pacing catheter previously removed from an adult Pace-Jector™ insertion device was sheathed within a 19 g epidural catheter, which had been shortened to allow easy passage of the pacing wire through its lumen. The pacing apparatus was threaded into the fetal right ventricle and the pigtail pacing wire advanced out of the sheath. The sheath and needle were removed, avoiding tension on the slack wire. The pacing wire was secured with suture to the maternal abdomen.

The pacing wire was attached to a Medtronic exernal pulse generator and ventricular pacing was initiated at a rate of 120/min. Ultrasound examination revealed ventricular contractions at 120/min. The pacemaker output was decreased until ventricular noncapture was apparent with return to a ventricular rate of 32. The ventricular capture threshold was 1.5 mA. The pacemaker was programmed to 6 mA ventricular output with a rate of 120 beats/min.

Course. The mother was sedated and placed at bed rest. External Doppler fetal heart tone monitoring revealed a heart rate of 120 beats/min. Four hours and 15 minutes after initiation of fetal pacing, intermittent irregular heart tones were noted. The pacemaker output was increased to 20 mA. Ventricular capture did not improve and within 5 minutes fetal heart sounds were no longer evident. Ultrasound examination revealed fetal asystole. The pacing catheter had not changed position and remained in the right ventricle. There was no hematoma in the ventricular wall and no thrombi within the ventricles. No change in the pericardial effusion was evident. The pacing catheter was removed and labor induced.

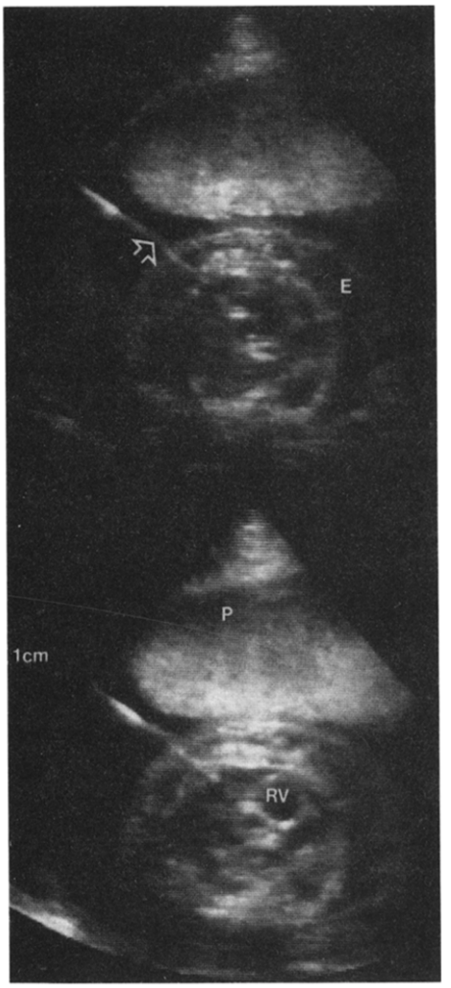

Figure 1. Two-dimensional fetal echocardiograms depicting the pacemaker lead passing through the chest wall and coiled in the right ventricle (RV). The **arrow** identifies the pacemaker cable; E = edema of chest wall; P = placenta.

Fetal autopsy. At postmortem examination, the heart was anatomically normal with right atrial and ventricular enlargement. The entry site in the right ventricle could not be identified. A serosanguinous pericardial effusion was present. Immunofluorescent studies were performed using labeled antibodies to human immunoglobulins G and A and C3 complement. Deposition of these three proteins was present in both ventricles.

Discussion

The application of transuterine-transthoracic fetal pacing for complete AV block has not been previously reported as a method of treatment for severe fetal bradyarrhythmia. Complete AV block may be a major cause of intrauterine fetal death in women with collagen vascular disease in whom the incidence of unexplained fetal death may exceed 10 percent (6). Litsey et al. (7) reported positive immunofluorescent studies of fetal atrial tissue which identified SSA (anti-Ro) antibodies and interruption of the specialized myocardial conduction fibers in the fetus. Identical findings were seen in our patient.

Medical management. There has been no adequate prenatal treatment for hydrops related to fetal bradycardia. When hydrops develops, the ventricular rate is usually less than 50 beats/min. Because this degree of bradycardia probably represents an idioventricular escape rhythm, it is unlikely that atropine would be effective. Catecholamines and beta-adrenergic stimulants may be tried but terbutaline and isoproterenol were not effective in this patient.

Goals of fetal pacing. The goal of fetal transthoracic ventricular pacing was to provide temporary acceleration of the fetal ventricular rate. In the absence of AV valve regurgitation, this should have improved cardiac output and allowed right atrial pressure to return to normal. Had pacing been effective in maintaining fetal life, digoxin would have been used to improve stroke volume and a diuretic agent used to enhance resolution of edema.

Limitations of fetal pacing. Fetal transthoracic pacing has several limitations. The present pacing lead is only moderately flexible and could become dislodged or cause injury with vigorous fetal movement. Fetal movements are minimal in severe hydrops, but fetal activity should increase with resolution of congestive heart failure. A longer lead with greater flexibility is required if prolonged temporary or implantable fetal pacemakers are to be developed. The external pacing system requires maternal bed rest and presents a risk of bacterial chorioamnionitis. Delivery of the fetus would require cesarean section to prevent dislodgment of the pacing catheter during delivery. The lead would need to be transected and pacing temporarily stopped during delivery. The placenta and fetus must be located in the uterus in positions that allow access to the heart. It is not known whether increasing ventricular rate rapidly with fetal pacing results in any metabolic or acid-base imbalances as a consequence of increasing myocardial oxygen consumption and cardiac output. In our patient, a slower acceleration of fetal cardiac rate might have prevented an adverse outcome.

Application of fetal pacing. Several factors appear favorable for future application of intrauterine pacing. The initial ventricular pacing threshold was extremely low in this hydropic fetus. If applicable to other fetuses, this would potentially allow the use of implantable fetal pacemakers. Sedation does not appear necessary to maintain inactivity of the hydropic fetus during placement of the pacing lead. No maternal or fetal morbidity could be directly attributed to insertion of the pacing lead. The bipolar pacing lead did not induce uterine contractions.

This technique requires the expertise of a team composed of a perinatologist, ultrasonographer and pediatric electrophysiologist. It should not be applied in the nonhydropic fetus unless physiologic data suggesting early congestive heart failure develop. Because the hydropic fetus is at such high risk for death in utero or if delivered, fetal pacing may be viewed as a reasonable extreme therapy. The development of a research program utilizing an animal model would be optimal before the application of this technique on a routine basis in the human fetus. Obstetrical contraindications should not be ignored in an attempt to save a hydropic fetus. However, despite these limitations, fetal pacing should be considered for the treatment of the dying fetus with complete AV block.

References

1. Kleinman CE, Donnerstein RL, Jaffe CC. Fetal echocardiography: a tool for evaluation of in utero cardiac arrhythmias and monitoring of in utero therapy: analysis of 71 patients. Am J Cardiol 1983;51:237–43.
2. Huhta JC, Strasburger JF, Carpenter RJ, Reiter A, Abinader E. Pulsed Doppler fetal echocardiography. J Clin Ultrasound 1985;13:247–54.
3. Kleinman CS, Donnerstein RL. Ultrasound assessment of cardiac function in the intact human fetus. J Am Coll Cardiol 1985;5:84S–94S.
4. Holsgreve W, Curry CJR, Golbus MS, Callen PW, Filly RA, Smith JC. Investigation of nonimmune hydrops fetalis. Am J Obstet Gynecol 1984;150:805–12.
5. DeVore GR, Siassi B, Platt LD. Fetal echocardiography. IV. M-mode assessment of ventricular size and contractility during the second and third trimesters of pregnancy in the normal fetus. Am J Obstet Gynecol 1984;150:981–8.
6. Gimovsky ML, Montoro M, Paul RH. Pregnancy outcome in women with systemic lupus erythematosus. Obstet Gynecol 1984;63:686–92.
7. Litsey SE, Noonan JA, O'Connor WN, Cottrill CM, Mitchell B. Maternal connective tissue disease and congenital heart block—demonstration of immunoglobulin in cardiac tissue. N Engl J Med 1985;312:98–100.